207* Recovery of lung function after intensive treatment due to pulmonary exacerbation in children with cystic fibrosis

R. Kraemer[1], S. Gallati[2], M.H. Schoeni[1]. [1]*University of Berne, Department of Paediatrics, Berne, Switzerland;* [2]*University of Berne, Division of Human Genetics, Berne, Switzerland*

Rationale: It is generally believed that lung function recovers after treatment of pulmonary exacerbations in CF, although it is unclear how often return to previous baseline is achieved.
Objectives: (1) Determination of the proportion of children who significantly improved or failed to recover lung function, and (2) identification of factors associated with success or failure.
Methods: Data from 24 CF-patients (15 males, 9 females) collected prior and after intensive treatment of pulmonary exacerbations, hospitalized on 44 occasions were investigated. Serial measurements pertaining to functional residual capacity (FRC_{pleth}, FRC_{MBNW}), lung clearance index (LCI), volume of trapped gas (V_{TG}), effective specific airway resistance (sR_{eff}), forced expiratory indices (FEV_1, FEF_{50}), PaO_2 and $PaCO_2$.
Results: Significant changes were observed for oxygenation given by PaO_2 ($p < 0.001$), as well as sR_{eff} ($p = 0.032$) and FEF_{50} ($p = 0.038$) indicating an improvement of lung function, however, only after 24 occasions (54.5%). After 13 occasions (29.5%) lung function remained within a ±8% change, and After 7 occasions (15.9%) lung function even deteriorated.
Conclusions: Improvement of oxygenation depended specifically on improvement of obstruction (sR_{eff}) and small airway function (FEF_{50}). In contrast, deterioration of gas exchange was closely associated with failure to recover ventilation inhomogeneities (LCI) and/or for the decrease of trapped gas (V_{TG}). In consequence, these complex physiopathological interactions regarding assessment of treatment efficacy demonstrate, that outcome parameters have to be carefully selected and defined, and not restricted to spirometric parameters only.

208* Long-term non-invasive ventilation in adults with cystic fibrosis: experience over two decades

W.G. Flight[1], J. Shaw[1], S. Johnson[1], K. Webb[1], A. Jones[1], A.M. Bentley[2], R. Bright-Thomas[1]. [1]*University Hospital of South Manchester, Manchester Adult Cystic Fibrosis Centre, Manchester, United Kingdom;* [2]*University Hospital of South Manchester, Long-Term Ventilation Service, Manchester, United Kingdom*

Objectives: Non-invasive ventilation (NIV) is accepted as a bridge to lung transplantation in CF but its role beyond this is unclear. We aimed to determine the effect of long-term NIV on lung function and survival in advanced CF.
Methods: We reviewed the records of all CF patients commenced on long-term NIV at the Manchester Adult CF Centre before October 2010. Weight, lung function and antibiotic therapy were recorded for the 3 years before and after initiation of NIV.
Results: 48 patients have received long-term NIV since 1991 and data were available for 47 of these. Median age at NIV set-up was 29 years (range 16–47). Mean±SD FEV_1%-predicted was 20.7±6.0%. 17 patients (36.2%) underwent lung transplantation, 13 (27.6%) died without transplantation, 3 (6.4%) had NIV withdrawn and 14 (29.8%) remain on NIV. Median time from NIV set-up to death or transplantation was 27 (3–90) and 16 (2–44) months respectively. Those still alive have been on NIV for a median of 14.5 (3–89) months. Mean IPAP & EPAP at set-up were 22.5 and 2.7cmH_2O.
FEV_1 and FVC fell by a mean of 398 and 685 ml over the 3 years before initiation of NIV but increased by 43 and 142 ml in the 3 years after NIV set-up. The improvements in FEV_1 and FVC at both 1 and 3 years post-NIV were statistically significant compared to equivalent periods before NIV ($p < 0.01$). There was no significant difference in IV antibiotic usage over the 12 months before and after NIV (median 65 v 86 days/yr; $p = 0.102$).
Conclusions: NIV appears to halt the decline in lung function in patients with advanced CF and improvements are maintained at three years of follow-up. This data supports the use of long-term NIV in advanced CF.

209 Inhaled hypertonic saline in infants with cystic fibrosis from Argentina

V. D'Alessandro[1], G. de la Fuente[1], G. Diez[1], E. Segal[1]. [1]*Children's Hospital, Pulmonary, La Plata, Argentina*

Introduction: Inhaled hypertonic saline (HS) using concentrations ranging from 3% to 7% has been used for sputum induction and positively affects lung function in children with cystic fibrosis (CF). However, safety and tolerability of HS in infants have been established in few studies.
Objective: To assess safety and tolerability of the inhalation of a single dose of 7% HS in infants with CF undergoing routine infant pulmonary function testing.
Material and Methids: Cross sectional study. We measured: Maximal Expiratory Flow at FRC ($V'max_{FRC}$) using rapid thoracic compression with Jaeger® equipment. The measurement was performed before and after the inhalation of 4 ml of 7% HS administered via face mask. Previous HS infants received four puffs of a short-acting bronchodilator. Infants had to be stable and free of acute respiratory infections. Oxygen saturation, respiratory rate, heart rate, cough, and wheezing were recorded throughout the study. T-test was performed to compare the means of the indicators.
Results: We evaluated 10 patients with a mean age of 18 months. Seven completed the entire study including HS lung function measurement. Mean value of $V'max_{FRC}$ did not exhibit a significant reduction from baseline to after HS inhalation values. Oxygen saturation, respiratory rate, heart rate did not change in any of the subjects during or after HS inhalation.
Conclusions: Results suggest that inhalation of a single dose HS is safe in infants with CF who are free of respiratory exacerbation and had pre-administration of a short bronchodilator prior saline inhalation. A longitudinal long term follow-up would also be helpful in order to evaluate the benefits of the treatment.

210 Dry powder inhalation with NaCl for increasing secretolysis in cystic fibrosis patients – a pilot study

U. Graepler-Mainka[1], V. Icheva[1], G. Herrmann[1], C. Adams[1], M. Stern[1], B. Zonnenberg[2], J. Riethmüller[3]. [1]*University Children's Hospital Tübingen, I, Tübingen, Germany;* [2]*Bartzon BV, Nieuwegein, Netherlands;* [3]*University Children's Hospital, Dept 2, Tuebingen, Germany*

Introduction: Inhaled hypertonic saline is standard care in CF-patients. Two work groups in Finland and Russia reported data about the halotherapy which applies dry powder saline in air in a special setting for patients with chronic lung disease. They found an increase in lung function parameters and a reduction of pulmonary exacerbations. In a small pilot study, halotherapy was proven for safety and efficacy in 6 CF-patients.
Methods: Dry powder aerosol was insufflated in a dry and well-tempered room with salt concentrations of 10 mg/m^3 in the air. Halotherapy was proven for 45 minutes on five consecutive days in a wellness area. Sputum was sampled, leukocytes and bacteria and lung function parameters (FEV1, MMEF) were analysed during, and one hour after, therapy. Secondary aims were side effects. No prophylactic antiobstructive therapy was implicated.
Results: We found in six adult CF-patients (i) a significantly increased secretolysis (mean amount of sputum 7.1±4.4g; $p = 0.008$) within and one hour after dry powder aerosol inhalation, (ii) a significant increase in leucocytes (mean 39±78/µl; $p = 0.049$) and *P. aeruginosa* colony counts (mean 0.3±0.5log; $p = 0.017$) in expectorated sputa and (iii) a significant improvement in lung function parameters, FEV1 (mean 3.8±2.5%; $p = 0.019$) and MMEF (mean 6.6±6%; $p = 0.05$) after 5 days of therapy.
Conclusion: These data confirm the findings of the trials carried out in Finland and Russia in patients with chronic lung diseases. We found a well-tolerated dry powder inhalation with an increased sputum production and similar clinical effects to saline inhalation in CF-patients, which must be consolidated by a larger clinical trial.